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Two distinct clinical types of interstitial lung disease associated with polymyositis-dermatomyositis

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Summary

Most patients with interstitial lung disease (ILD) associated with collagen vascular diseases (CVD) have a chronic indolent course with a relatively favorable prognosis; however, acute progression has been reported in some polymyositis-dermatomyositis patients. This study evaluated the prevalence, clinical features, and outcome relative to the presentation type of ILD in polymyositis-dermatomyositis (PM-DM).

Ninety-nine patients with newly diagnosed polymyositis-dermatomyositis seen at the Asan Medical Center in Korea between January 1990 and December 2004 were enrolled. The clinical, radiological, and pathological findings were retrospectively reviewed. ILD were divided into acute (dyspnea within 1 month before diagnosis) or chronic types.

ILD was found on chest radiographs in 33 patients (33.3%), and 11 (33.3%) of these were considered acute. The acute group presented with more severe respiratory symptoms, hypoxemia, and poorer lung function. Patients with an acute presentation had ground glass opacity and consolidation on high-resolution computed tomography (HRCT), in contrast to reticulation and honeycombing in the chronic type. Surgical lung biopsy of one acute-type patient revealed diffuse alveolar damage, whereas biopsies in the chronic type showed usual interstitial pneumonia (UIP) in four cases and nonspecific interstitial pneumonia (NSIP) in another four. Eight acute-type patients (72.7%) died of respiratory failure within 1–2 months despite steroid therapy. The 3-year mortality rate of the chronic-type patients

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(21.2%) was not statistically significantly different from that of the patients without ILD (10.2%).

In polymyositis-dermatomyositis, the acute, severe form of ILD was not infrequent.

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Introduction

Interstitial lung disease (ILD) is frequent in collagen vascular diseases (CVD) such as scleroderma, Sjögren's syndrome, systemic lupus erythematosus, rheumatoid arthritis, and polymyositis-dermatomyositis (PM-DM).¹⁻⁷ Although the clinical features of ILD associated with CVD are similar to those of idiopathic interstitial pneumonia (IIP), the survival in CVD-ILD is much better, and most patients have a chronic indolent course.⁸⁻¹¹ Nevertheless, there is a subset of patients with PM-DM who have rapidly progressive respiratory failure, and this subset has high mortality compared with the overall survival of PM-DM-ILD.¹²⁻¹⁴ Consequently, the survival curve of PM-DM shows an initial rapid drop in the first year after diagnosis, representing the accelerated mortality of this subset, and a slower decline thereafter.^{15,16}

A review of surgical lung biopsy specimens in cases of ILD associated with scleroderma and Sjögren's syndrome has revealed that the nonspecific interstitial pneumonia (NSIP) pattern is more common than "usual interstitial pneumonia" (UIP).^{2,5,17,18} Similarly, in PM-DM, Douglas et al.^{19,20} and Cottin et al.^{19,20} reported that NSIP was the most common pattern. However, in Douglas et al.^{19,20} surgical lung biopsies were reviewed in only 31% of the patients. Therefore, the reported surgical lung biopsy results might not provide the entire picture for PM-DM-ILD. At our institution, almost all patients with PM-DM admitted for a diagnostic work-up routinely have a chest radiograph, so it is relatively easy to check for the presence of ILD. This study reviews the overall clinical features of a relatively large series of patients with PM-DM, focusing on the relative prevalence of ILD in this group, to determine the clinical and radiological findings of the acute type of ILD.

Patients and methods

Study population

This retrospective study was performed at Asan Medical Center, a 2000 bed, university affiliated, tertiary referral center in Seoul, Korea. From January 1990 to December 2004, 107 patients were newly diagnosed with PM-DM based on Bohan and Peter's criteria: symmetric proximal muscle weakness, with or without dysphasia or respiratory muscle weakness; muscle enzyme elevation; electromyographic abnormalities; compatible muscle biopsy; and skin rash of dermatomyositis.^{21,22} Eight patients with overlap syndrome combined with other CVDs were excluded, and thus 99 patients were included in the study. All the patients had a chest radiograph taken at the time of diagnosis, and interstitial pneumonia was found in 33 patients (Table 1). The interstitial pneumonia was divided into two groups according to the respiratory symptoms. The acute type of ILD (acute ILD) was defined as the development of rapidly

progressive severe dyspnea and hypoxemia requiring oxygen therapy or ventilator care within one month preceding diagnosis,²³⁻²⁷ and the others were classified as the chronic type (chronic ILD).

Clinical and laboratory findings

Clinical data and laboratory results were extracted from the patients' medical records. The factors examined included clinical history, physical examination, laboratory tests, pulmonary function tests, radiological findings, bronchoalveolar lavage (BAL) fluid findings, and biopsy results.

Chest radiographs and computed tomography (CT) of the chest

Chest radiographs and high-resolution computed tomography (HRCT) examinations of the lungs were reviewed and interpreted by a single-chest radiologist blinded to the biopsy results and clinical outcomes. Radiological evidence of ILD on chest X-ray includes ground glass opacity, patchy bilateral consolidation, and basal-predominant reticular opacities. HRCT was performed for the differential diagnosis of ILD in patients with respiratory symptoms or abnormal chest radiographs. Five patients with respiratory symptoms and normal chest radiographs underwent HRCT. Two of them had a normal HRCT, and the remaining three proved to have an infection. Another 16 patients with abnormal chest radiographs were excluded because of heart failure ($n = 5$), infection ($n = 9$), carcinoid tumor, or amyloidosis with bilateral radiological infiltration. The radiologist (J.B.S.) determined the presence, degree, and extent of consolidation, ground glass opacity (GGO), traction bronchiectasis, reticulation, and honeycombing on HRCT.²⁸

Table 1 Definition for interstitial lung disease (ILD).

Interstitial lung disease associated with PM-DM

- (1) Symptoms/signs: dry cough, exertional dyspnea, and crackles at both lung bases
- (2) Chest radiograph or HRCT (bilateral reticular/reticulonodular infiltration, ground glass opacity, airspace consolidation, or honeycombing)
- (3) PFT: restrictive defect or low diffusion capacity
- (4) No evidence of known causes, such as infection, heart failure, and drug reaction
- (5) Surgical lung biopsies

Pulmonary function tests (PFTs)

Spirometry was performed with a Sensor Medics 2100 (Yorba Linda, CA, USA), the diffusion capacity of the lung for carbon monoxide (DL_{CO}) was measured with a Sensor Medics Model V_{max} 22, and total lung capacity (TLC) was measured using a Sensor Medics Auto Box 6200. All results are expressed as a percentage of normal predicted values,²⁹ and values below 80% of normal were considered abnormal. Improvement or worsening was defined as a change greater than 10% of the forced vital capacity (FVC) or TLC (more than 15% in DL_{CO}) in either direction.³⁰

Surgical lung biopsy

A surgical lung biopsy was performed to confirm the pattern of interstitial pneumonia. Given the patients' general condition, a surgical lung biopsy was performed in nine patients only. Two lung pathologists (M.K. and T.V.C.) reviewed the lung biopsy slides independently, and the histology of each lung was classified according to the patterns defined in the new ATS/European Respiratory Society consensus classification for IIPs.³¹

Statistical analysis

The data are presented as the mean \pm SD for continuous variables and as percentages for categorical variables. Statistical analyses were performed using SPSS for Windows version 11.0 (Chicago, IL). Continuous variables were compared using the Mann-Whitney *U* test or the Kruskal-Wallis test, whereas categorical variables were compared using the chi-square test or Fisher's exact test. Cumulative survival probabilities were estimated using the Kaplan-Meier method. The log-rank test was used to compare the survival of groups of patients. A *p* value < 0.05 was defined as statistically significant (all tests were two tailed).

Results

Prevalence of ILD by chest radiography

ILD was diagnosed in 33 of the 99 patients (33.3%) by chest X-ray. Of the 33 patients with ILD, 11 (33.3%) were acute, and 22 (66.7%) were chronic type. ILD and PM-DM were diagnosed simultaneously in 22 of the 33 patients (66.7%), and ILD was diagnosed before PM-DM in six patients (18.2%).

Clinical features and laboratory findings at the time of diagnosis

The patient demographics, symptoms, laboratory tests, and BAL fluid findings recorded at initial presentation are summarized in Table 2. The PM-DM patients were divided into three groups, depending on the presence and type of ILD. Inflammatory markers such as the erythrocyte sedimentation rate (ESR) and serum albumin level differed among the three groups ($p < 0.05$); there were no significant differences in the demographic features or muscle enzymes. Antinuclear antibody (ANA) had a higher prevalence in the

PM-DM patients without ILD. A positive ANA reaction might be a good prognostic marker (odds ratio 0.258, 95% confidence interval 0.060–1.105, $p = 0.068$). Jo-1 antibody had a higher prevalence in the chronic ILD group. In the patients with acute ILD, the respiratory symptoms were not only more severe but were also the main reason for admission in the majority (73%) of the patients. By contrast, most of the patients with chronic ILD had only minimal respiratory symptoms, and about one-third were asymptomatic. Of those without ILD, 14 patients complained of dyspnea, and further work-up, such as 2-D echocardiography, exercise PFTs, measured maximal inspiratory pressure, HRCT, and lung biopsy, revealed the causes of the dyspnea to be heart failure ($n = 5$), combined pneumonia ($n = 4$), muscle weakness ($n = 3$), cystic bronchiectasis ($n = 1$), and amyloidosis ($n = 1$). The BAL fluid in the chronic ILD cases showed lymphocytosis, in contrast to the elevated percentage of neutrophils in the acute-type cases, although the difference was not statistically significant due to the small number of patients who underwent BAL.

PFTs

PFTs were performed in 28 of the 33 patients with ILD and in 18 of the 66 patients without ILD (Table 3). Five patients (45.5%) with acute ILD could not complete the PFTs because of severe dyspnea and hypoxemia. The lung function of the remaining six patients with acute ILD was still the lowest as compared with the other groups. Among the patients with ILD, a restrictive defect (92.9%) was the most common lung function abnormality, followed by a low DL_{CO} (88.0%), and 80% of the patients had a restrictive defect with a low DL_{CO} . Among the patients without ILD, three had a mildly restrictive defect attributable to respiratory muscle weakness, and another four patients had a restrictive pattern with a low DL_{CO} attributable to concomitant diseases, such as heart failure ($n = 2$), pneumonia ($n = 1$), and bronchiectasis ($n = 1$).

HRCT of the chest

HRCT was performed in 31 of the 33 patients in the ILD group. The major findings are summarized in Table 4. In acute ILD, all the patients had consolidation ($n = 8$) and GGO ($n = 2$), whereas in chronic ILD, reticular densities with ($n = 3$) or without honeycombing predominated (Fig. 1). The most common distribution of the abnormalities on HRCT was basal and subpleural in both groups. Five patients with acute ILD had a predominantly peribronchial distribution.

Surgical lung biopsies

Surgical lung biopsies were performed in nine patients only (eight chronic and one acute ILD). Many patients with acute ILD were too ill to undergo a surgical lung biopsy and started treatment with high-dose steroids shortly after admission. The pathologic findings in the nine biopsied cases were an organizing diffuse alveolar damage (DAD) pattern in the single acute ILD case and NSIP (4 cases) and UIP (4 cases) in the chronic ILD cases.

Table 2 Summary of the initial clinico-laboratory data for the patients with polymyositis-dermatomyositis.*

	Without ILD	With ILD		<i>p</i> Value [†]
		Acute type	Chronic type	
Number of patients	66	11	22	
Male/female, Number	29/37	4/7	10/12	NS
Age, years	47.2 ± 16.0	47.9 ± 4.1	48.9 ± 2.7	NS
Smoking history, Number				
Never/former/current	44/6/13	9/0/1	12/1/8	NS
<i>Symptoms and signs at the time of admission, %</i>				
Dyspnea as a chief complaint	81.8	31.8	0.004	
Cough	9.1	100.0	45.5	0.000
Dyspnea	21.2	81.8	59.1	0.000
Fever	25.8	45.5	45.5	NS
<i>Laboratory tests</i>				
PF ratio mm Hg		231 ± 147	385 ± 82	0.004
ESR, mm/h	35.9 ± 30.9	56.2 ± 29.3	53.1 ± 25.9	0.027
LDH, IU/L	1324 ± 999	2604 ± 2511	1613 ± 253	NS
CK, IU/L	3787 ± 6111	1054 ± 1954	3012 ± 3199	NS
Aldolase, SU/mL	16.5 ± 17.1	18.4 ± 16.8	24.3 ± 25.1	NS
Albumin, g/dL	3.4 ± 0.1	2.9 ± 0.1	2.9 ± 0.2	0.000
Antinuclear antibodies (ANA)	22/66	0/11	4/22	0.010
Positive/negative, no. (%)	(33.3%)	(0%)	(18.2%)	
Anti-Jo-1 Positive/negative, No.	1/38	1/8	3/13	0.040
<i>Initial BAL finding</i>				
Macrophages, %		55.8 ± 35.0	36.9 ± 27.1	NS
Lymphocytes, %		12.1 ± 10.6	47.4 ± 21.9	0.032
Neutrophils, %		29.1 ± 41.0	13.9 ± 10.6	NS
Eosinophils, %		2.9 ± 4.5	1.6 ± 3.0	NS

PF ratio: PaO₂/FiO₂ ratio.

*Data are presented as the mean ± SD unless indicated otherwise.

[†]Exact test for categorical variables and Mann-Whitney *U* test or Kruskal-Wallis test for continuous variables.**Table 3** Results of the initial pulmonary function tests in patients with interstitial lung disease and polymyositis-dermatomyositis.*

	PM-DM without ILD	PM-DM with ILD	<i>p</i> Value
Number of patients	18	28	
FVC (% predicted)	78.3 ± 14.9	60.5 ± 13.7	0.001
FEV ₁ (% predicted)	85.4 ± 18.0	67.2 ± 13.8	0.001
FEV ₁ /FVC (%)	81.8 ± 11.0	86.9 ± 10.4	NS
DL _{CO} (% predicted)	80.6 ± 21.2	58.1 ± 15.7	0.005
Number of patients		17	
TLC (% predicted) [†]		84.0 ± 30.8	

*Data are presented as the mean ± SD.

[†]TLC data of patients without ILD were not available.**Table 4** Initial high-resolution computed tomography and histopathological findings.

	Acute ILD	Chronic ILD
<i>Major pattern in the initial HRCT</i>		
Number of patients	10	21
Consolidation	8	1
Ground glass opacity (GGO)	2	1
Reticulation		16
Honeycombing		3
<i>Histological findings</i>		
Number of patients	1	8
DAD	1	
NSIP		4
UIP		4

Clinical course of ILD

Acute type of ILD

Eight of the 11 patients (72.7%) with acute ILD died of rapidly progressing respiratory failure within 1–2 months

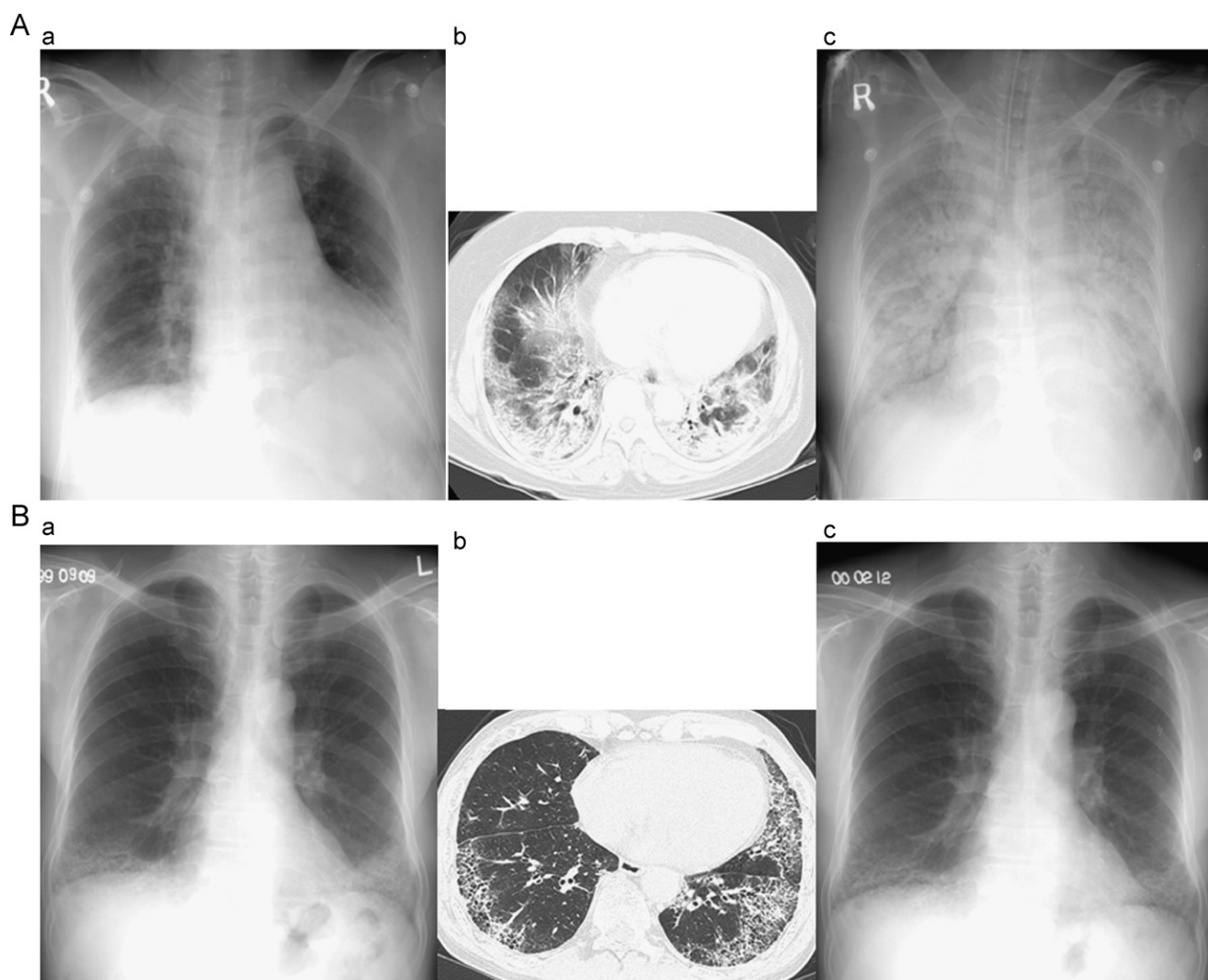


Figure 1 Radiological findings of the patients with acute and chronic interstitial lung disease. (A) Acute ILD. (a) The chest X-ray taken at the initial presentation shows slightly increased interstitial densities in both lower lung fields, and (b) HRCT shows a subpleural distribution of GGO with focal consolidation. (c) The chest X-ray taken 2 weeks later shows increased opacities throughout the entire lung fields with an endotracheal tube. (B) Chronic ILD. (a) The chest X-ray taken at the initial presentation shows slightly increased interstitial densities in both lower lung fields, and (b) HR-CT shows a subpleural distribution of reticular densities with traction bronchiectasis. (c) The chest X-ray taken 5 months later shows no significant change compared with the initial chest X-ray.

despite all therapeutic endeavors, including steroid pulse therapy and immunosuppressant therapy. Non-survivors had a lower oxygen tension than survivors, but there were no differences in the demographic, clinical, or laboratory findings between the survivors and non-survivors (Table 5). Although the numbers were too small for statistical comparison, the extent at HRCT seemed to be more widespread in the non-survivors. Three survivors had a patchier peribronchial distribution, suggesting a bronchiolitis obliterans organizing pneumonia (BOOP) pattern. Three survivors were stable or partially improved clinically after 3, 5, and 9 years, respectively.

Chronic type of ILD

All 22 patients were treated with prednisolone either alone (22.7%) or in combination with immunosuppressants (77.3%).

Six months after the diagnosis, lung function had improved in the survivors compared with the initial values (FVC: 67.7 ± 20.2 vs. $60.7 \pm 13.7\%$ predicted, $p = 0.034$; DL_{CO} : 63.7 ± 19.2 vs. $58.9 \pm 16.5\%$ predicted, $p = 0.093$). On long term follow-up, PFTs revealed the maintenance of the improved state for up to 3 years (Fig. 2). In terms of myositis, 11 patients (50%) exhibited complete remission, and eight (36.3%) showed partial improvement in muscle strength.

Survival

As a whole, the patients with ILD had significantly lower survival rates than those without ILD. The 3-year survival rate of the patients with ILD was 61.6% and that of the patients without ILD was 89.8% ($p < 0.05$, Fig. 3). Subgroup analysis of the patients with ILD showed that the 3-year

Table 5 Comparison of survivors and non-survivors in acute interstitial lung disease.*

	Non-survivor	Survivor	P value
Number of patients	8	3	
Male/female gender, Number	4/4	0/3	0.068
Age, years	47.6 ± 15.2	48.7 ± 10.1	0.776
PF ratio, mm Hg	187 ± 76	348 ± 245	0.376
PaCO ₂ , mm Hg	31.6 ± 8.6	36.5 ± 3.9	0.376
WBC, /mm ³	7700 ± 4300	12100 ± 9000	0.376
ESR, mm/h	59.9 ± 33.4	47.7 ± 19.4	0.667
CK, IU/L	1424 ± 3026	2061 ± 2950	0.921
Aldolase, SU/mL	22.1 ± 40.0	32.0 ± 18.8	0.250
Major Pattern on HRCT, Number			
Consolidation	5	3	0.467
Ground glass opacity	2		
Surgical lung biopsy	DAD(1)		

*Data are presented as the mean ± SD, unless indicated otherwise.

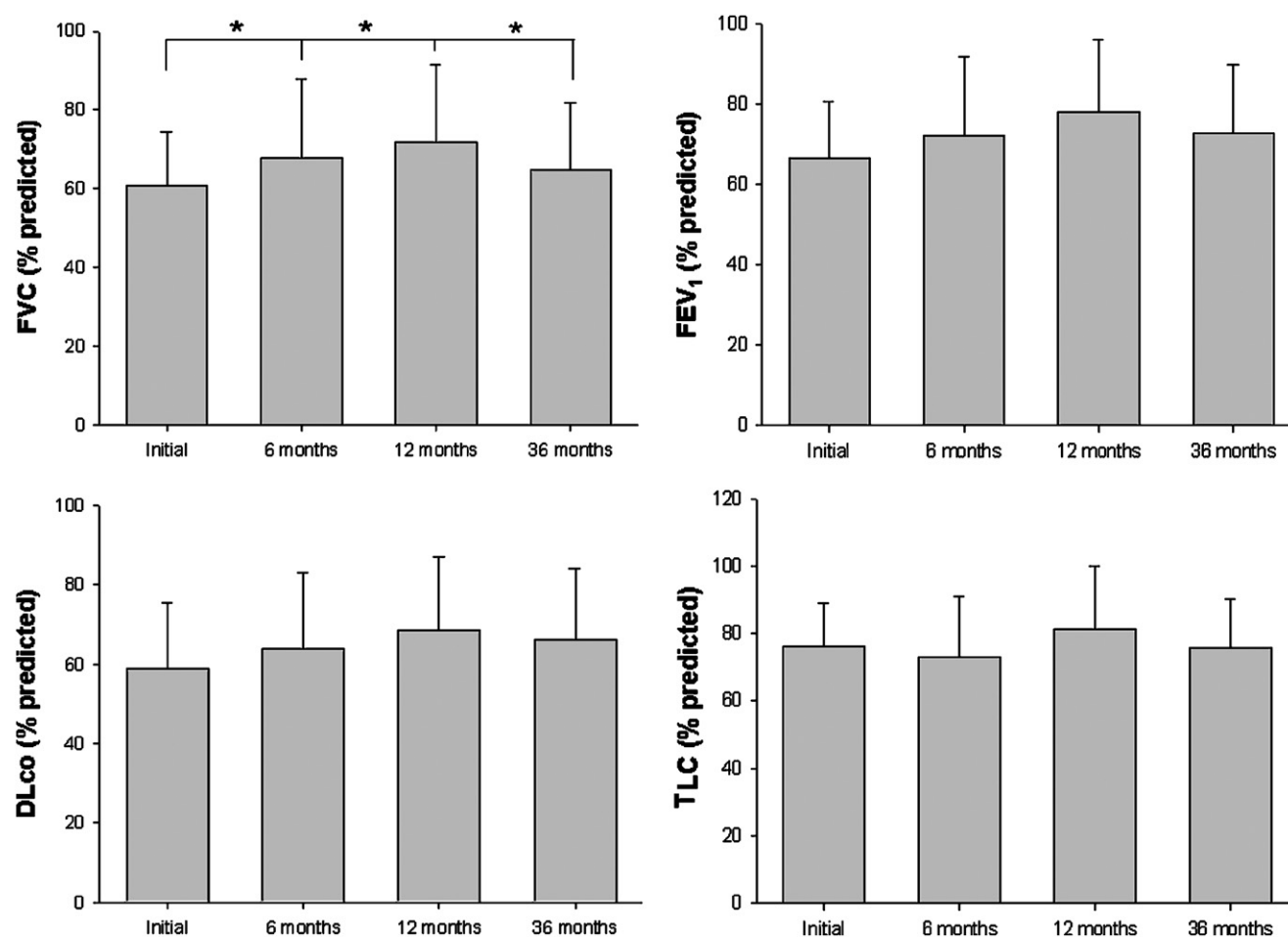


Figure 2 Changes in pulmonary function of the patients with chronic type of interstitial lung disease*. *The data are presented as the mean ± SD. $p < 0.05$ compared with initial pulmonary functions.

survival rate was 27.3% for the acute form and 78.8% for the chronic form ($p < 0.05$). The survival rate of the chronic ILD group was not significantly different from that of the patients without ILD.

Both the survival rate and the cause of death differed between the two ILD groups. Eight patients died of progressive ILD in the acute ILD group, whereas the causes of death in the chronic-ILD group were sepsis with multi-

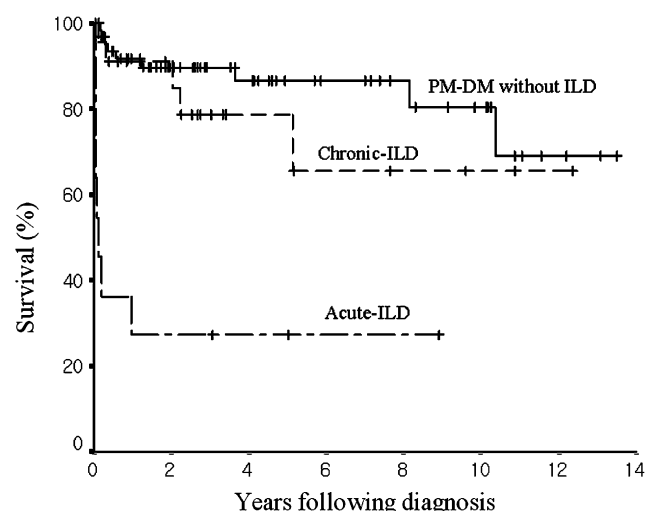


Figure 3 Overall survival curves of the patients with polymyositis-dermatomyositis (PM-DM) according to the state of lung involvement ($p < 0.05$).

organ failure ($n = 3$), superimposed pneumonia ($n = 1$), and cancer progression ($n = 1$).

The causes of death in the PM-DM patients without ILD included sepsis ($n = 4$), cancer progression ($n = 3$), superimposed pneumonia ($n = 1$), and heart failure ($n = 1$).

Discussion

Here, we have shown that the clinical type of ILD associated with PM-DM differs from the chronic course of other CVD-ILDs.^{6,8} One-third of the patients with ILD associated with PM-DM had the acute severe form, with rapidly progressive respiratory failure and high mortality. An acute form of ILD is encountered in other CVDs, but it is relatively rare.^{32,33} As almost all newly diagnosed PM-DM patients at our institution were admitted for a diagnostic work-up, we believe that our estimation of the prevalence of ILD in this group of patients approach to the true prevalence. Furthermore, detailed histories with laboratory and radiological findings were available, providing overall clinical pictures of ILD in PM-DM.

Most CVDs present with a variety of pleuropulmonary manifestations, with ILD being one of the most common. The reported prevalence of ILD in CVDs varies not only with the nature of the CVD but also the method of detection.^{6,34–36} The prevalence of ILD in PM-DM patients has been reported to be in the range of 5–46%, depending on the method used. In most studies, the presence of ILD affected the treatment and outcome of PM-DM.^{23,37,38} In our study, one-third of the patients had ILD on chest X-ray at presentation; this might actually be an underestimation of the true prevalence because of the relative insensitivity of chest radiography. However, we think that this includes all the patients with clinically significant disease, given that 24% of the patients with radiological ILD were still asymptomatic and that further work-up of other patients complaining of respiratory symptoms failed to reveal more patients with ILD.

Many reports have suggested that the prognosis of CVD-ILD is much better than that of IIP, despite their clinical and

radiological similarities.^{8–11} The ATS/ERS Consensus Classification of IIPs subclassifies IIPs into seven different types, and many studies have reported that idiopathic NSIP has a better prognosis than IPF/UIP.^{39–46} A recent review of surgical lung biopsy specimens of CVD-ILD revealed that the NSIP pattern was predominant in most CVDs, such as scleroderma, PM-DM, and Sjögren's syndrome.^{2,5,17–20} In contrast, the UIP pattern is predominant in IIP.⁴⁷ It is uncertain whether the better survival in CVD-ILD is attributable mainly to the relative frequency of NSIP with its more favorable prognosis or whether there is a genuine biological difference between IIP and CVD-ILD. Similar to the diversity of CVDs, the forms of ILD in the various CVDs differ considerably in their frequency. For example, in scleroderma, most reports agreed that the NSIP pattern is predominant.^{17,18} However, in Sjögren's syndrome, although the NSIP pattern is the most frequent, many different kinds of ILD have been reported, including UIP, LIP, amyloidosis, and even lymphoma.^{2,28,48} In rheumatoid arthritis, not only different types of interstitial pneumonia but also small airway diseases are not infrequent, and UIP is reported to be more common than the NSIP pattern.^{3,49} Regardless of the histological pattern, most cases of CVD-ILD seem to follow a relatively chronic indolent course,^{6–8} and there are only isolated case reports of acute forms of ILD in CVD other than PM-DM.^{32,33} By contrast, there are many reports of acute ILD in PM-DM, although the relative prevalence or detailed clinical comparisons (versus chronic disease) are lacking.^{12,13,50–52} The reports that showed the predominance of the NSIP pattern in PM-DM may result from selection bias because surgical lung biopsies tend to be performed in relatively stable patients.^{19,20}

Thus, we studied the relative prevalence and clinical characteristics of the subtypes of ILD in PM-DM and confirmed that ILD in PM-DM can be divided into two clinical types with different radiological and pathological findings and different outcomes. Furthermore, our results revealed that acute ILD is not infrequent (33% of the ILD associated PM-DM). Many patients are too ill to tolerate either surgical lung biopsies or pulmonary function tests. Most of them were admitted for respiratory symptoms with severe hypoxemia, rather than muscle problems, suggesting that clinicians should be alert to the possibility of PM-DM in this situation. Most died of rapidly progressing respiratory failure within 1–2 months, despite high-dose steroid therapy and ventilator care. Only one patient had a surgical lung biopsy, which revealed a DAD pattern similar to other reports.^{12–14} A BOOP pattern may also have a clinical presentation similar to a DAD pattern, as reported in PM-DM.^{13,53} Unfortunately, we cannot confirm the relative prevalence of the BOOP and DAD patterns in PM-DM. Patients with the BOOP pattern have been reported to have a better prognosis than those with UIP or DAD patterns, not only in IIP but also in CVD-ILD.^{13,33,40,41} The three patients with the acute-type ILD who survived in our series had HRCT features more suggestive of BOOP, i.e. bilateral multifocal consolidation in the bronchial distribution, but the diagnoses could not be confirmed histologically.

The chronic form of ILD in PM-DM, which is more common than the acute type, presented with slowly progressive dyspnea and cough, with many cases being asymptomatic. HRCT revealed reticular densities with or without

honeycombing in most of the patients and subpleural GGO in some of them. Three patients had honeycombing on the HRCT at the time of the diagnosis of PM-DM, had developed respiratory symptoms 6–24 months prior to the diagnosis of PM-DM. It has been well known that in some patients, the ILD can precede the manifestation of CVD for several years and long standing ILD represented by honeycombing was reported in other studies.^{6,7,19,54–58}

Owing to the small number of the surgical lung biopsies, we could not confirm the predominance of the NSIP pattern in chronic-type ILD in PM-DM. The course of chronic ILD was stable or improved with treatment in most patients on long term follow-up. Survival analysis showed that these patients with chronic ILD had a survival rate similar to that of the patients without ILD.

The reported characteristics of acute ILD include the lesser complaints of muscle weakness, a lack of elevation of serum creatine phosphokinase, and negative tests for autoantibodies (anti-Jo-1 antibodies).^{59,60} However, we could not confirm all of these findings. The higher ESR and lower albumin level in our patients with ILD suggest that patients with ILD have more severe systemic inflammation than those without ILD.

The mortality of PM-DM patients has declined with early diagnosis and improved treatment modalities.^{61,62} Many deaths in these patients result from respiratory failure, including those who develop progressive ILD and pneumonia or sepsis. A shorter disease history, rash, dysphagia, fever, older age, and leukocytosis have been associated with reduced survival.¹⁵ In our study, acute ILD had a high mortality rate, requiring more aggressive treatment with a combination of high dose steroids, immunosuppressive agents, and newer modalities, as in other reports.^{50,63} This study has several potential limitations. First, the frequency of ILD might be underestimated due to the comparatively low sensitivity of chest radiography. Second, a pathologic diagnosis of ILD was made in only a few patients. Third, the prevalence of acute ILD might have been overestimated, as our clinic is a university-affiliated, tertiary referral center.

As a conclusion, we showed that the acute severe form of ILD is not infrequent (33% of the ILD associated PM-DM), unlike the predominance of the chronic form of ILD observed in other CVDs. This acute type presented with respiratory symptoms rather than muscle problems, and HRCT might be useful for predicting the prognosis.

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